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Case report

Small-cell carcinoma of the kidney: Report of a case and review of the literature

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ABSTRACT

Extrapulmonary small-cell carcinoma (EPSCC) is a rare small-cell carcinoma (SCC). It is recognized as a clinicopathological entity distinct from SCCs of the tracheobronchial tree. EPSCCs account for about 2.5% of all SCCs. SCCs of the kidney are an even rarer neoplasm. Abdominal pain and hematuria are the major clinical symptoms. Many cases are already advanced disease at diagnosis and have poor prognoses. Platinum-based chemotherapy seems to be beneficial for overall survival. We describe an 82-year-old male patient who was seen at the emergency room due to complaints of gross hematuria and right-flank pain. Computed tomography showed a large right renal tumor. Treatment modalities, pathologic features, survival outcome of this case, and a review of similar cases in the literature are presented and discussed. To the best of our knowledge, this is the 21st case of SCC of the kidney reported in the literature to date. It is also the first report in Taiwan of a primary SCC of the kidney with the tumor thrombus extending to the inferior vena cava which required a right radical nephrectomy.

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1. Introduction

Extrapulmonary small-cell carcinoma (EPSCC), although a rare neoplasm, is now being increasingly reported. EPSCC was first described in 1930 by Duguid and Kennedy.¹ It accounts for about 2.5% of all small-cell carcinomas (SCCs).² The primary site of occurrence was reported in a variety of organs, such as the head and neck region, gastrointestinal tract, genitourinary tract, and others.^{2–5} The most common site in the genitourinary tract is the urinary bladder. Primary SCC of the kidney is an extremely rare neoplasm with aggressive behavior. Many of these cases had advanced disease at diagnosis and have a poor prognosis. We report a case of SCC of the kidney and reviewed the literature on this rare neoplasm.

2. Case report

This 82-year-old male patient had good general health and denied any previous systemic disease. He was a non-smoker. No body weight loss had been noted in the past year. However, painless gross hematuria had been noted for 1 year. In addition, he had suffered from right-flank and lower-abdominal-quadrant pain for 1 month. He visited the emergency room, where abdominal

sonography revealed a right renal mass. Computed tomography (CT) disclosed a heterogeneous contrast-enhanced mass occupying the upper pole of the right kidney.

During admission, a physical examination revealed a right-flank mass. Urinalysis showed hemopyuria [red blood cell count >100/high-power field (HPF) and white blood cell count >100/HPF]. A hemogram showed mild anemia (hemoglobin of 10.3 g/dL). His preoperative creatinine level was normal (1.2 mg/dL). No active lung lesion was noted on a chest film. Retrograde pyelography was done, but the right renal pelvis could not be demonstrated due to difficulty with the contrast medium entering this area. A CT scan revealed a 7.3 × 6.0-cm heterogeneous contrast-enhanced mass in the upper pole of the right kidney. A tumor thrombus in the inferior vena cava and clusters of enlarged lymph nodes in the retrocaval and para-aortic spaces and right renal hilum were also demonstrated on the CT scan (Fig. 1). A whole-body bone scan showed no evidence of bone metastasis.

Under the impression of a right renal tumor, he received a transperitoneal laparoscopic radical nephrectomy. The operative course was smooth with blood loss of about 50 mL. The operative time was 4 hours. During the operation, multiple enlarged lymph nodes were noted around the renal hilar area with severe adhesion to the inferior renal cava. Complete lymph-node dissection was too difficult and too dangerous to perform. The pathology report revealed SCC with angiolymphatic invasion. A tumor thrombus in the renal vein was also noted. Histopathologic pictures showed small, oval to spindle-shaped tumor cells. A higher magnification revealed tumor cells with hyperchromatic nuclei and scanty

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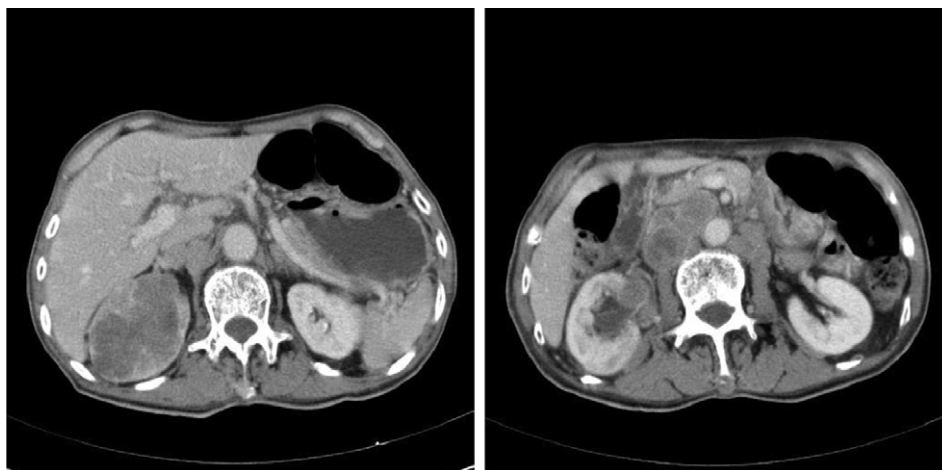


Fig. 1. Computed tomography scan showing a 7.3 × 6.0-cm heterogeneous contrast-enhanced mass in the upper pole of the right kidney (left) with right para-aortic and retrocaval lymph nodes and an inferior vena cava tumor thrombus (right).

cytoplasm after hematoxylin and eosin staining (Fig. 2). Immunohistochemical studies revealed positive reactivity for cytokeratin, neuron-specific enolase (NSE), synaptophysin (Fig. 3), and chromogranin A (Fig. 4). The final pathologic stage was T3aN1M0, stage IV. He received a Port-a-Cath (Smith Medical, PORT-A-CATH II Low Profile) implantation and was then referred to the oncology department for further adjuvant chemotherapy. The patient received four courses of chemotherapy with starting doses of 300 mg/m² carboplatin and 75 mg/m² etoposide. Unfortunately, follow-up CT revealed multiple nodal metastases in the para-aortocaval space, peritoneal space, and bilateral common iliac chain 4 months postoperatively. The general condition of the patient gradually worsened, and he was referred to hospice care. He died 9 months after the laparoscopic radical nephrectomy.

3. Discussion

EPSCC is a rare neoplasm. It accounts for approximately 2.5% of all SCCs. The first case report of an EPSEC that occurred in the mediastinal gland was in 1930.¹ In the US, the overall incidence of EPSCC is estimated to be 0.1–0.4% of all cancers.⁶

EPSCC can occur in various organs including those of the gastrointestinal and genitourinary tracts. The most common site of

occurrence in the genitourinary tract is the urinary bladder.³ Primary SCC of the kidney is an extremely rare disease. Only 20 cases of SCC of the kidney have been reported in the literature to date.⁷ According to Majhail's report, the medium age at diagnosis was 62 years with a female predominance (male:female ratio of 1:3.4).²

Clinical symptoms of a renal SCC are indistinguishable from those of a renal cell carcinoma. Abdominal pain and hematuria are the most common clinical features.^{1–3} The behavior of this rare neoplasm seems quite aggressive. Early dissemination and frequent recurrence are common. About 32% of patients showed distant metastasis at the time of diagnosis.^{1,3}

Imaging studies, including chest X-ray, and chest and abdominal CT scans, should be carried out, but it is still difficult to distinguish an SCC from a renal cell carcinoma. Magnetic resonance imaging of an EPSCC shows a diminished signal on T1-weighted images and a heterogeneous mixed signal on T2-weighted images. In addition, the predominant medullary location and lack of central necrosis are features of EPSCC, which are unusual for a renal cell carcinoma.⁸ A whole-body bone scan should also be done to detect early dissemination.

The differential diagnosis of EPSCC includes renal cell carcinoma, urothelial carcinoma, renal sarcomas, and other metastatic

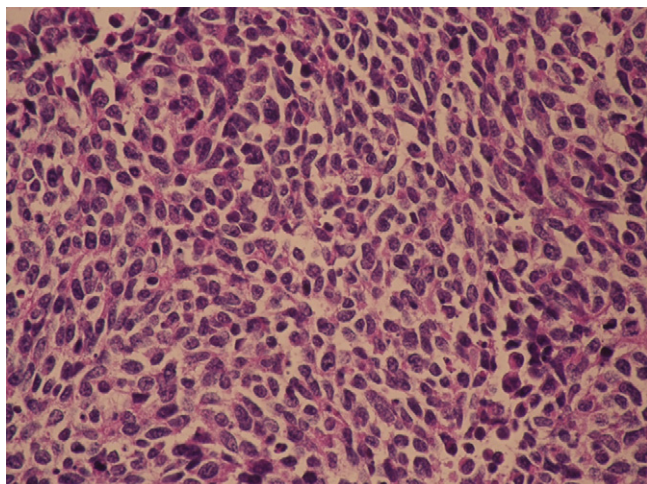


Fig. 2. Tumor cells with hyperchromatic nuclei, indistinct nucleoli, and scanty cytoplasm (hematoxylin and eosin stain; 400×).

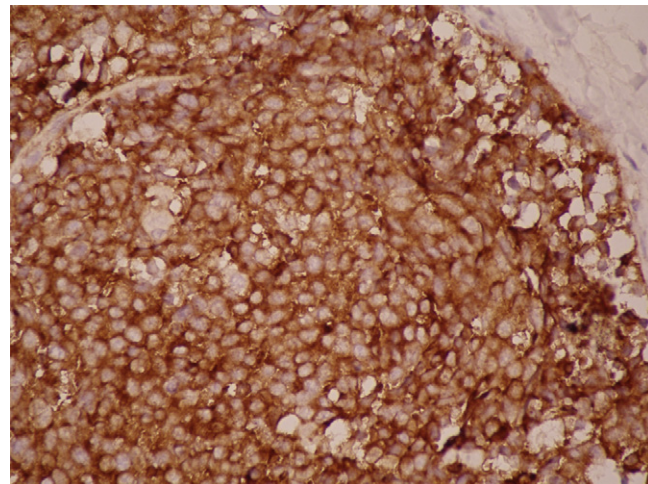


Fig. 3. Immunohistochemical studies showing diffuse positive staining for synaptophysin (original magnification, 400×).

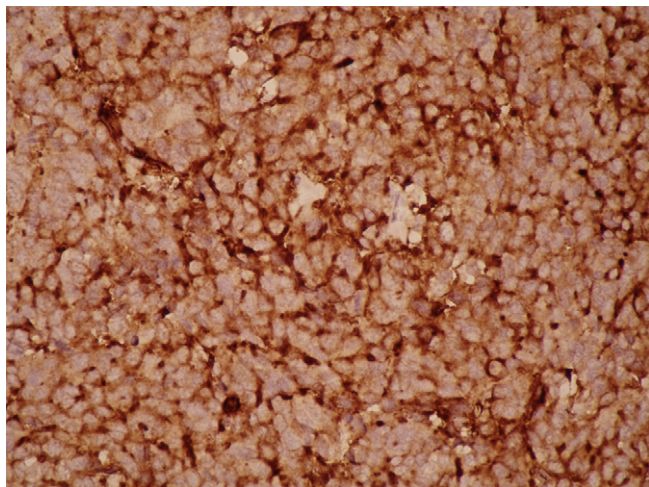


Fig. 4. Immunostaining showing diffuse positive staining for chromogranin A (original magnification, 400×).

tumors. The final diagnosis depends on the histopathological characteristics, such as the appearance of round to spindle-shaped small cells with dense nuclei, inconspicuous nucleoli, and sparse cytoplasm. For equivocal cases, immunohistochemical staining with chromogranin A, synaptophysin, or NSE may be helpful.^{1,3}

In the treatment of EPSCC, surgery alone is not enough. In the published literature, a combination of surgical resection and chemotherapy offered better results.^{1–3} Due to the local extensiveness and early dissemination of this rare tumor,

chemotherapy can be of some help. The combination of platinum-based chemotherapy and etoposide, 5-fluorouracil, or gemcitabine may improve overall survival. The median overall survival was 8 months in non-chemotherapy patients, and 20 months in patients receiving platinum-based combination chemotherapy ($p = 0.02$).¹

In conclusion, SCC of the kidney is an extremely rare neoplasm with aggressive behavior and a high propensity for locoregional dissemination. The clinical presentation is usually late with a poor prognosis. Combination chemotherapy with a platinum-based regimen showed benefits against tumor progression and for overall survival.

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